CASE REPORT

Radicular Myoclonus in a Patient with Guillain-Barré Syndrome

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Abstract

A 53-year-old Japanese woman presented with myoclonus during the course of Guillain-Barré syndrome. The myoclonus was characterized by relatively regular involuntary movements, starting from proximal muscles of the right lower leg, and moving almost simultaneously towards the left lower leg and upper trunk. Surface electromyography revealed rhythmic synchronous discharges with 100-200 ms duration in the agonist and antagonist muscles at approximately 4 Hz. The jerk-locked back averaging, long latency reflexes, and somatosensory evoked potentials studies were normal. We report myoclonus due to radiculitis in a patient with Guillain-Barré syndrome.

Key words: Guillain-Barré syndrome, radicular myoclonus, radiculitis, demyelination, ephaptic transmission


Introduction

Guillain-Barré syndrome (GBS) is an acute inflammatory disorder of the peripheral nervous system. There have been only a few reports of GBS-associated involuntary movements including myokimia, opsoclonus, and ballism (1-6). However, myoclonus has not been reported in patients with GBS. Here, we describe a patient with GBS that presented with myoclonus probably due to severe radiculitis.

Case Report

A 53-year-old Japanese woman with no history of neuropsychiatric illnesses visited our outpatient clinic due to muscle weakness. She did not have preceding upper respiratory inflammation or diarrhea.

Neurological examination showed proximally dominant muscle weakness in the four limbs, the absence of tendon reflexes, and anesthesia with glove and stocking type distribution. Nerve conduction studies (NCS) revealed prolonged distal latencies, for example 8.8 ms in the right median nerve, and absent F waves on all examined nerves. Cerebrospinal fluid (CSF) examination showed no cells but elevation of protein (200 mg/dL). No antiganglioside antibodies were detected in the serum. Contrast-enhanced MRI studies disclosed enhancement in the posterior roots at thoracic and lumbar levels of the spinal cord. GBS was diagnosed according to the criteria proposed by Asbury and Cornblath (7). We administrated immunoglobulin intravenously (0.4 g/kg/day for 5 days) from day 1. However, her weakness progressed to tetraparesis, and bilateral abducens and bilateral facial palsy became evident. On day 4, respiratory failure developed, and then she was intubated. Follow-up NCSs demonstrated marked prolongation of distal latency (21.0 ms in the right median nerve) on day 16. Protein of CSF increased to 695 mg/dL on day 8. On day 17, CSF became coagulated soon after the spinal tap. On day 19, she displayed almost rhythmic involuntary movements in the proximal limb muscles and trunk with variable amplitude between oscillations. The involuntary movements always started from proximal muscles of the right lower leg, and moved almost simultaneously towards the left lower leg and upper trunk, lasting for more than 10 minutes. Furthermore, the movements occurred spontaneously, but they were often elicited by patellar tendon reflex. Surface electromyography (EMG) revealed rhythmic synchronous discharges with 100-200 ms duration in the agonist and antagonist muscles at ap-
proximately 4 Hz (Fig). Electroencephalography (EEG) was normal. Back averaging of EEGs time-locked to the onset of myoclonic discharges demonstrated the absence of preceding cortical spikes or premovement potentials at Cz, C3, and C4. No long latency reflexes (LLR) were evoked during tonic activation by stimulation of the accessory nerve. Somatosensory evoked potentials (SEP) showed no extremely enhanced cortical components. Auditory brainstem responses were normal. This involuntary movement was slightly relieved by Clonazepam, but not by distraction, psychotherapy, or placebo. The involuntary movements dramatically disappeared with improvement of muscle weakness and decrease of CSF protein.

**Discussion**

Myoclonus is defined as a sudden brief jerk caused by involuntary muscle activity; it is classified into cortical, subcortical, and spinal (8, 9). However, myoclonus of peripheral origin including the spinal roots has also been reported (10, 11).

The involuntary movement in the present patient was characterized by rhythmic synchronous discharges with 100-200 ms duration in the agonist and antagonist muscles at approximately 4 Hz on surface EMG. These characteristics corresponded to rhythmic myoclonus rather than tremor. The jerk-locked back averaging, LLR, and SEP studies did not support that the movement originated from the cortex. Subcortical myoclonus, such as reticular reflex myoclonus and dystonic myoclonus, often appears in muscles innervated by cranial nerves. The myoclonus in the current patient consistently started from the right lower leg. The distribution pattern of our patient was different from that of subcortical myoclonus. Thus, these findings suggest that the origin of the myoclonus could be neither the cortex nor subcortex. The MR evidence of enhancement of spinal roots, together with no abnormal signals in the cortex, brainstem, or spinal cord, indicates that the patient had severe radiculitis. Pathogenesis in GBS is generally acknowledged as inflammatory demyelination of the spinal roots with perivascular and endoneurial infiltrates of lymphocytes and macrophages (12); radiculitis is also known to cause spinal myoclonus-like movement disorder (13). Furthermore, the disappearance of the myoclonus with improvement of muscle weakness and decrease in CSF protein also reinforces the hypothesis that radiculitis related to GBS might be the focus of the myoclonus. Hence, the myoclonus in the present patient is considered to be radicular myoclonus resulting from GBS-associated radiculitis.

Ephaptic transmission is considered to be a possible pathophysiology in myoclonus. Smith et al. showed spontaneous generation of compound action potentials propagated along demyelinating dorsal roots in an animal study (14). In the present patient, the myoclonus occurred spontaneously and moved up rostally. Dorsal roots are thought to be involved in our patient and cause self cross-excitation of a large population of neurons through ephaptic transmission.

Tremor has been reported in peripheral neuropathies including chronic inflammatory demyelinating neuropathy (CIDP) (15, 16). Dalakas et al. reported that 7 patients with CIDP showed positional tremor prominent in the distal muscles of the upper extremities. Tremor varied in frequency between 3.3 and 6.4 Hz among the patients with CIDP (16). The frequency of myoclonus in our patient was similar to those of the patients with CIDP. However, tremor in the present patient was prominent in the proximal limb muscles and trunk, indicating that the generator of myoclonus in our patient was different from that of the patients with CIDP.

In conclusion, we report myoclonus due to radiculitis in a patient with GBS. GBS can cause radicular myoclonus.

The authors state that they have no Conflict of Interest (COI).

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**References**


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